East African Scholars Journal of Medicine and Surgery
Abbreviated Key Title: EAS J Med Surg
ISSN: 2663-1857 (Print) & ISSN: 2663-7332 (Online)
Published By East African Scholars Publisher, Kenya

Volume-4 | Issue-8 | Aug-2022 | DOI: 10.36349/easjms.2022.v04i08.002

OPEN ACCESS

Case Report

Survival of Haemophilic Patient in Surgery Case Report

Olivia M Kimario[1*], Aron Kiberiti[1], James Komanya[1], Sr Alice Masenga[2], Alex Donasiano[1]

[1]Department of otorhinolaryngology, Catholic University of Health and Allied Sciences, P.O. Box 1464, Mwanza Tanzania
[2]Department of Paediatric Surgery, Catholic University of Health and Allied Sciences, P.O. Box 1464, Mwanza Tanzania

Article History
Received: 13.06.2022
Accepted: 12.07.2022
Published: 04.08.2022

Journal homepage:
https://www.easpublisher.com

Quick Response Code

Abstract: Haemophilia is an inherited bleeding disorder. The blood does not clot as it should, which can result in spontaneous bleeding and bruising after surgery or other injuries. Hemophilia A is an X-linked, recessive disorder caused by deficiency of functional plasma clotting factor VIII, which may be inherited or arise from spontaneous mutation. Hemophilia B, is an X-linked inherited bleeding disorder, usually manifested in males and transmitted by females who carry the causative mutation on the X chromosome is a deficiency of factor 1X. We are presenting a case of 34years old man with the complain of bilateral neck swelling for 5days,difficult in swallowing and breathing for 3days.Emergency tracheostomy was done and admitted to the ICU thereafter CT-scan was done and showed retropharyngeal swelling. The excision of the mass was done whereby hematoma was evacuated. Patient was admitted to the ICU for about two weeks then transfer to the wards. The laboratory test revealed deficiency of factor V111.

Keywords: Haemophilia, inherited bleeding disorder, spontaneous mutation .

INTRODUCTION

Haemophilia is a disease that results due to coagulation disorder. There are various groups of hemophilia that is group A,B, C and parahaemophilia. Haemophilia A is a genetic bleeding disorder associated with the deficiency of clotting factor VIII, Haemophilia B is genetically associated with deficiency of factor 1X,hemophilia C deficiency of factor X1 and parahemophilia deficiency of factor V [1-3]. These factors deficiency can be inherited or acquired and have different levels of severity [4, 5].

All hereditary coagulation factors' deficiencies are autosomal recessive, so they can manifest in both gender but Haemophilia A and B are X-linked disorders(3). Females can rarely been affected. In contrast all other coagulation factors deficiencies are autosomal recessive [6- 8].

The prevalence of Haemophilia A (FVIII deficiency) is 1 in 5000 to 10,000 while haemophilia B affects about 1 in 40,000 males at birth. Factor XI deficiency is also called hemophilia C and Rosenthal syndrome. It was first recognized in 1953 in patients who experienced severe bleeding after dental extractions [10]. Considered a rare disorder, the estimated prevalence is 1 in 1,000,000. In populations where consanguinity is common, higher prevalence has been reported (8% among Ashkenazi Jews of Israel).

An important differential consideration is an accurate drug history, if available, from the patient's card (record), especially because of new oral anticoagulants. These drugs cannot be monitored by the usual laboratory tests. Autoimmune diseases, malignancies or pregnancies might indicate an acquired haemophilia [11].

In minor forms of haemophilia, the administration of desmopressin is required to limit the bleeding signs [12, 13]. Patients affected with the moderate form, who rarely have bleeding, are substituted (with spontaneous bleeding or prior to surgery) with factor VIII. Patients with severe haemophilia and frequent bleeding events receive long-term prophylactic therapy with the administration of factor VIII three times per week [12].

At the beginning (during the first 10 days or more), a level of 100% of FVIII:C should be achieved. The administration of 1 IE/kg body weight results in a factor VIII:C increase of approximately 2%. The therapy should be continued in low doses as a maintenance therapy because the half-life of factor VIII is 12 h [11]. Laboratory monitoring of factor VIII and maintenance of a level of 100% should be continued in severe injury patients until all invasive procedures are completed.

***Corresponding Author:** Olivia Michael Kimario
Depatment of ENT CUHAS P.O. Box 1464 Mwanza, Tanzania

CASE PRESENTATİON

A 34 year old male came to our emergency department with complains of throat pain, bilateral neck swelling for 5days, difficulty in swallowing, and difficulty in breathing for 3 days. This complains were of gradually onset and as time goes by was getting worse. The difficult in swallowing was for both solid and liquid food both; Due to the continuous increase in size of the swelling it lead to the difficult of breathing and swallowing. Also the mother explains of the bleeding tendency in the family and elaborates of difficulties of blood to stop whenever had a slightly bruise or a mirror injury; explains of the fact she heard an idea that this is a problem; she has not gone to the hospitals to analyze what it is. No history of blood transfusion reported or any surgery.

On general examination patient was dyspnea with stable vitals. Local examination: bilateral neck swelling equal in size measures about 6cm by 8cm,firm and slightly tender, mobile not fixed to underline structures.

We had a diagnosis of retropharyngeal tumor. Immediately tracheostomy was done to alleviate the patient symptoms. Patient was taken to the ICU due to continuous bleeding tendencies from surgical site were tracheostomy was done and was kept on Mechanical ventilation. When was at ICU several investigations were done.FBP results showed high neutrophils, PT and INR normal but the APTT was high 61.4(25.5-39.6).Radiological imaging which was CT scan showed retropharyngeal mass bulging from both sides of the neck.

He was transferred to ward and received transfusion of 2units of whole blood and one FFP. but in HDU for close minitoring of vitals, Tracheostomy and bleeding. Patient stayed in the wards for 3 day then we did trasoral surgery. Where by evacuated a lot of hematoma then packed the patient. Due to his bleeding tendencies patient was sent to the ICU. In the ICU kept the patient on tranesmic acid 500mg iv for 5days, Ampiclox 500mg iv tds 3/7, PCM 1g iv tds 2/7To receive 3units of whole blood and 2 FFP. Patient was out of the ICU after 2weeks then transferd to HDU. Therefore blood sample taken for lab work was out with the bleeding disorders which was of Hemophilia A which is deficiency of factor 8 the ; After initial Therapy for the haemophilia condition patient was then decannulated from the tracheostomy and discharged and is currently under hematology clinic with follow up every few weeks.

DISCUSSION

Congenital hemophilia A is an X-linked recessive coagulation disorder caused by factor VIII deficiency. This disorder is characterized by a bleeding tendency, a normal PT, and a prolonged aPTT. Also as the most common congenital coagulopathy, hemophilia A occurs commonly in male births. As our patients presented with the same characteristics and has factor V111 as analyses done in the laboratory. Female sex is rarely involved [6]. In severe form of hemophilia, deep bleeding such as intra-articular and intramuscular hemorrhage is seen in infancy and early childhood due to increased mobility and activity and unusual purpura or hematoma may also trigger the diagnosis.

In the management of haemophilia with surgical condition is done with fast and accurate treatment for survival. Most of the times unusual bleeding episodes, tendencies to develop haematomas, epistaxis or even muscle and joint bleeding after low-impact trauma in the family and the patient's history should be explicitly questioned. Prior to primary haemostasis, which is not impaired in haemophilia, excessive bleeding from the smallest cut or abrasions is not present [12]. As in this case this patient has not been having frequent episodes of been admitted due to bleeding episodes so its likely that had been impacting trauma which is of low impact that results to hematoma that brings about upper airway obstruction.

Management of this disease condition in our resource limit isn`t easy as sometimes blood and fresh frozen plasma may not be available or not enough. As to this patient in the initial management received two unit with one FFP in the second procedure received 3 units of whole blood and 2FFP. This show if have deficiency of blood this can not be attempted. Also the laboratory test to identify the deficiency factor may not be available at certain time. Its a challenge to put everything at hand in a resource limited countries.

CONCLUSION

Managing a patient with bleeding disorder isn`t easy but this should not let us leave them alone instead should take courage and help them out.

Authors Contributions
OMK: Writing – original draft
AK: Conceptualization
Conceptualization

Declaration of conflicts of interest: The authors declare no conflicts of interest.

Consent: The informed consent was obtained from the patient in this case report

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Cite This Article: Olivia M Kimario, Aron Kiberiti, James Komanya, Sr Alice Masenga & Alex Donasiano (2022). Survival of Haemophilic Patient in Surgery Case Report. *East African Scholars J Med Surg, 4*(8), 167-169.